
Children's Special Health Internal Policy/Statement Cystic Fibrosis (CF)

Description

Cystic Fibrosis (CF) is an inherited multi-system disease, particularly affecting the lungs and digestive system. CF is caused by a defect in a gene that makes a protein which controls the movement of salt and water in and out of the cells. This causes thick, sticky secretions in the respiratory, digestive tract, and reproductive system. It also causes increased salt in sweat on the skin

Diagnostic Criteria

- Newborn metabolic screening results
- Sweat chloride test (standard test for diagnosing CF)
- DNA
- Pancreatic and lung function tests

CSH Coverage

- Only **providers** listed on the Eligibility Letter will be paid
- Labs/Tests must be performed by a Wyoming Medicaid provider
- Well Child Checks (coverage limited to Pediatrician) according to AAP Periodicity Schedule
- Medications
 - Antibiotics
 - Replacement enzymes
 - Bronchodilators
 - Steroids
- Equipment/Supplies
 - Therapy vests (for chest percussion to loosen mucus from lungs)

Contact CSH for questions regarding additional medications and/or equipment/supplies

Minimum Standards of Care/Care Coordination

Refer to Care Coordination Manual, Ch. 3, Pg. 8, Child and Family Assessment

- Perform **Nursing Assessment** with detailed focus on the following:
 - Respiratory system (i.e. cough, sputum, shortness of breath, chest congestion)
 - Exercise and physical activity tolerance
 - Gastrointestinal system (i.e. nutrition, appetite, weight loss, bleeding, intestinal blockage, blood sugar)
 - Genital/Urinary system (i.e. poor urine output, nephritis, rectal prolapse, sexual maturation, bowel movements)
 - Muscular-skeletal system (i.e. short stature, clubbing)
 - Current medications/any side effects or reactions
 - Known food and/or drug allergies
 - Height and weight, plot on growth curve
- Encourage testing as recommended by the American Academy of Pediatrics (AAP)
- Encourage pneumococcal and influenza vaccinations
- School performance and behavior
- Encourage family and child to live as "normal and active" life as possible

Contact CSH if family is Non-Compliant (i.e. repeated missed appointments, failure to follow healthcare plan)

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• **Referrals** that may be recommended (CSH prefers Pediatric Specialist, if possible)

Visits to Providers may be limited due to budget

- Pulmonary Specialist
- Gastroenterologist
- Urologist
- Otolaryngologist (ENT)
- Geneticist
- Cardiologist
- Mental Health
- Dietician
- Link the child and family with appropriate and needed services

Specialists may or may not be covered by CSH Program

Well Child Checks

- Immunizations (including vaccinations)
- Assess and follow-up any abnormal findings
- Dental
- Vision
- Hearing

• Emergency Preparedness Plan

- Medic Alert ID bracelet/necklace should be encouraged
- Medical Emergency Plan of what to do for the child's care when away from home or with a different caregiver (i.e. chest percussion, respiratory distress, GI bleed, rectal prolapse)
- Discuss self-management of the disease
- Encourage the family to speak with the child's school in regards to the school's policy on Cystic Fibrosis and emergency plan (i.e. salt depletion)

• Health Record

- Encourage family to maintain a record of the child's health information ("Packaging Wisdom" as a suggestion) that includes:
 - Medication administration
 - Type
 - Dosage/Frequency, any side effects or response to medication
 - Respiratory status
 - Frequency/Duration of cough
 - Changes in sputum
 - Gastrointestinal status
 - Weight loss/gain
 - Exercise tolerance
 - Joint pain
 - Treatment/procedures and hospitalizations
 - List of providers and contact information, if available

Transition

Refer to the Care Coordination Manual, Ch. 3, Pg. 10, Coordinating Care

- Discuss with the family if the child is eligible for an IFSP, IEP, or qualify for Section 504 according to the American Disability Act (ADA)
- Encourage family to do life-care planning (i.e. medical expenses, physical limitations, sexuality)
- Support and encourage psychosocial considerations such as:
 - Dependence-Independence issues
 - Risky behavior (i.e. substance abuse, sexual acts)
 - Educational and Vocational planning

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